Section: Anaesthesiology

Case Report

NAVIGATING ANAESTHETIC CHALLENGES IN A CHILD WITH ARTHROGRYPOSIS MULTIPLEX CONGENITA : A CASE REPORT

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Abstract

Arthrogryposis multiplex congenita (AMC) is a rare congenital syndrome characterized by multiple joint contractures and associated with various congenital anomalies. So, appropriate preoperative evaluation is mandatory to rule out congenital anomalies. Anaesthetic management is challenging in these patients because of difficult airway, difficult intravenous access and difficult positioning. Here we report a case of 3 year old female child with congenital dislocation of left hip posted for varus derotation osteotomy under general anaesthesia and caudal epidural block.

INTRODUCTION

AMC is a rare non progressive congenital syndrome with an incidence of 1/3000 live birth.[1] It is characterized by multiple joint contractures and fibrosis with an intact sensory system and associated with multiple congenital anomalies like cleft palate, genitourinary defects, cardiac defects, pulmonary hypoplasia and vertebral anomalies.[2,3] These patients can have difficult airway because of micrognathia, high arched palate, short, rigid and webbed neck.[4] These patients also have difficult intravenous access because of tense skin and minimal subcutaneous tissue. These patients often requires anaesthetic exposure for the correction of orthopedic deformities and their perioperative care can be challenging for anaesthesiologists.

CASE REPORT

Figure 1: (A) contracture of upper limb with thumb in palm deformity; (B) intraoperative picture showing micrognathia and EJV cannulation.

A 3 year old, 10 kg female child with a known case of AMC diagnosed at birth. The child was having congenital dislocation of left hip and was posted for varus derotation osteotomy. Preoperative assessment was done a day prior to the surgery. There was history of preterm cessarean delivery at 32 week POG in view of breech presentation. The patient was having birth weight of 1 kg and was having history of NICU stay for 15 days and also received phototherapy for neonatal jaundice. There was history of surgery for bilateral CTEV under general anaesthesia. There was history of 1 unit packed cell volume transfusion two months back. There was no history of pneumonia, seizures, cyanosis, drug allergy and recent upper respiratory tract infection. Patient was immunised till date. Airway examination revealed mallampati grading III, micrognathia and slight buck teeth. The patient was having normal neck movements. On chest auscultation, air entry was equal on both sides. The haematological and biochemical investigations were within the normal range. Written and informed consent was taken from the parents following the discussion on the possibility of the difficult airway.
The child was fasted for 6 hours for solids and 2 hours for liquids prior to surgery and premedicated with syrup phenargan 5 mg a night before and 2 hours prior to the surgery. After identification and checking the consent, the patient was shifted on the table. Due to anticipated difficult airway, a difficult airway cart was kept ready. The expected surgical duration was 4 hours. Standard ASA monitoring was conducted. Baseline vital parameters showed a heart rate of 120/min, oxygen saturation of 98% on room air and blood pressure of 90/65 mmHg. Preoxygenation with

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100% oxygen started and under sevoflurane induction, external jugular vein cannulation was done after failed peripheral intravenous access. Injection glycopyrrolate in a dose of 0.005 mg/kg and fentanyl in a dose of 2 mcg/kg was given. Main aim during induction of anaesthesia was preservation of spontaneous ventilation, which was achieved through ketamine given in a dose of 2 mg/kg. Following loss of consciousness, patient deepened using sevoflurane to achieve adequate plane of anaesthesia for laryngoscopy. Check laryngoscopy was performed after optimal positioning and Cormack Lehane grading was found to be 3b and on giving backward upward rightward pressure improved to 2b and patient was intubated using OETT of internal diameter 4.5 mm with the help of stylet in 2nd attempt. Subsequently a loading dose of 0.5 mg/kg and maintenance dose of 0.1 mg/kg of injection atracurium was administered throughout the surgery. Anaesthesia was maintained with N2O, O2 and sevoflurane to achieve minimum alveolar concentration of 1. For intraoperative analgesia, caudal epidural block using 5 ml of 0.25% bupivacaine was given. After completion of surgery, reversal using injection neostigmine in a dose of 0.05 mg/kg and injection glycopyrrolate in a dose of 0.01 mg/kg was given and patient was extubated once protective airway reflexes and adequate muscle power returned. Patient was shifted to recovery room and after 1 hour of observation shifted to orthopaedic ward.

DISCUSSION

AMC is usually associated with multiple congenital anomalies, such as musculoskeletal deformities, genitourinary defects, congenital heart disease, pulmonary hypoplasia, vertebral anomalies etc.[2] The primary pathology is in-utero restriction of fetal joint mobility resulting in fixation of joint and growth of extra connective tissue around a functionally normal joint resulting in further restriction of joint movement.[5] There are various syndromes associated with it including Freeman Sheldan syndrome, Brown syndrome, Bruck syndrome, ARC syndrome etc.[6-9] There are multiple challenges faced by anaesthesiologists including difficult airway, difficult intravenous access, difficult positioning, cardiorespiratory problems, malignant hyperthermia and difficult regional blockade.

AMC patients have difficult airway accounting for various maxillofacial findings like micrognathia, high arched palate and decreased mandibular opening.[4] In our patient also, we anticipated difficult airway because of micrognathia and mallampati grading of 3. So, difficult airway cart was kept ready and patient was induced with ketamine and sevoflurane with the maintenance of spontaneous ventilation. There is also the risk of malignant hyperthermia intraoperatively and perioperatively, although the association is controversial between AMC and MH. Temperature monitoring can be done intraoperatively. In patients with underlying myopathy, there may be hyperkalemic response with use of depolarising muscle relaxant.[10] So, we avoided succinylcholine despite predicted difficult airway. As our patient had previous history of 2 surgeries under general anaesthesia, So we used inhalational induction during securing an intravenous accesss and maintenance of anaesthesia.

Reduced subcutaneous tissue and tense and glossy skin accounts for difficult intravenous access in these patients. We also encountered difficult intravenous access and had to cannulate the external jugular vein. Intraoperative positioning is difficult to maintain due to multiple contracture of extremities. So, proper padding should be done on bony prominences to avoid compression pressure.

The use of regional anaesthesia remains a topic of debate because one of the etiologies of AMC is a self-limited anterior horn cell disease. There are various case reports which employed successful use of caudal epidural block for management of postoperative pain in Freeman Sheldon syndrome which is a variant of AMC. Fascia iliaca block was successfully used for muscle biopsy in AMC.[11,12] We also successfully used, caudal epidural block for intraoperative analgesia which reduce the dose of opioids.

CONCLUSION

Perioperative care in AMC is challenging because of possibilities of difficult airway, difficult intravenous access, difficult positioning and difficult regional blockade. Careful preoperative evaluation should be carried out to rule out congenital anomalies. Careful airway evaluation, perioperative monitoring of body temperature, tachycardia and hypercarbia are needed to avoid critical events.

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